Telemedicine system for a FSHD family with chronic respiratory failure

Portaro S.[1], Bramanti A.[2], B. Alagna[1], Bramanti P.[1], Lunetta C.[3]

[1]IRCCS "Bonino-Pulejo" Research institute, SS 113, c.da Casazza, 98124, Messina, Italy
[2]Institute of applied sciences and intelligent systems «E. Caianello» (ISASI) – National research council of Italy (CNR), Messina, Italy
[3]Centro Clinico Nemo, Milan, Italy

Background

- ✓ Facioscapulohumeral muscular dystrophy (FSHD) is an autosomal dominant inherited disorder, characterized by asymmetric involvement of muscles in the facial, upper extremity, trunk and lower extremity region with variable severity [1].
- ✓ Although respiratory weakness is a relatively unknown feature of FSHD, it is not uncommon [1].
- ✓ Telemedicine has been used in a variety of health care fields, but only recently, with the advent of sophisticated technology, its interest among health professionals became evident [2].
- ✓ The aim of this study is to describe the utility of a telemedicine system based on videoconferencing and telemonitoring of cardiorespiratory variables for the telecare of 4 siblings with FSHD and chronic respiratory failure. In fact, the peculiarities presented by the severe forms of FSHD make it advisable to design and evaluate telemedicine systems that meet their needs

Patients & Methods

- ✓ The present study involved four siblings affected by a severe form of FSHD (FSHD score 15/15), wheelchair bound, with chronic respiratory failure and long-term mechanical ventilation, living in a rural area far away from the referral centre for neuromuscular diseases (Fig.1)
- ✓ All siblings received a 6-months period of telemedicine support
- ✓ The telemedicine system was based on videoconferencing regarding:
- ❑ Neurological assessment – once a month and on-call h24 : the neurologist coordinate medical interventions and managed the emergencies.
- ❑ Psychological support: every two weeks

Furthermore, we adminstered to all patients the following instruments:

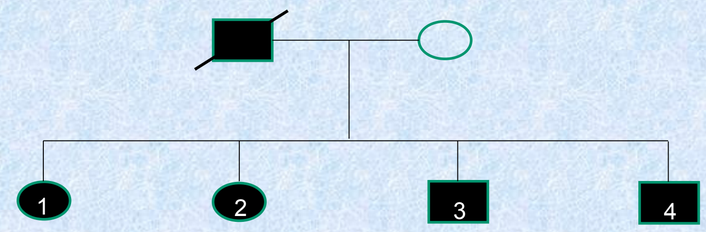
- The 36-Item Short Form Survey (SF 36) to investigate changes in quality of life in eight fields (Physical Functioning, Limitations Due To Phisical Problems, Limitations due to Emotional Problems, Pain, General Health, Vitality, Social Functioning , Mental Health);
- The Hospital Anxiety and Depression Scale (HADS) to evaluate mood and emotional state;
- The Psychosocial Impact of Assistive Devices Scale (PIADS) to verify the effect of telemedicine service on some Psychosocial features as ability, adaptability and self-esteem.

We also administered a questionnaire to caregivers, the Caregiver Burden Inventory, to evaluate levels of perceived burden.

All instruments, except for PIADS, were administered before using telemedicine service (T0) and after six months (T1).

- ❑ Nurse-coach monitoring for devices' use – performed daily
- ❑ Telemonitoring of cardiorespiratory variables (oxygen saturation and heart rate) – performed daily and if needed, in relation to patients' symptoms

Fig. 1 . Family tree



Results

- ✓ Overall we performed 540 videoconference sessions per patient, including:
  - a daily contact with short monitoring oximetry measurements and heart-rate measurements
  - psychological support – We found a mild improvement in mood and emotional status of patients. Only one patient showed significant reduction of depression and anxiety level by HADS while the remaining three patients reported positive improvement in "mental health" subdimension of SF36 at T1. Regarding the caregivers, telemedicine service has not produced changes in burden's level. In addition, PIADS' scores revealed higher average scores in "ability", that is a good skill to face problems (Table 1).
  - Only one episode of desaturation occurred, managed by the neruologist, supporting the general practitioner who was present during the acute event
  - nurse-coach monitoring for devices' use was performed daily
- ✓ With respect to the clinical impact, after enrolment in the telemedicine program, the total number of hospital admissions for acute events fell.

Table 1

CBI

Subject	T0	T1
Caregiver 1	26	26
Caregiver 2	10	10
Caregiver 3	18	18

HADS scale

Patient	DepressionT0	Depression T1	Anxiety T0	Anxiety T1
1	16	11	10	6
2	7	7	7	5
3	5	5	11	9
4	1	1	4	4

SF36 – T0 scale

Patient	Self-esteem	Adaptability	Ability
1	0,87	0,83	0,58
2	0,62	1,16	0,91
3	0,37	0,5	1,08
4	0,37	0,16	0,25

SF36 – T1 scale

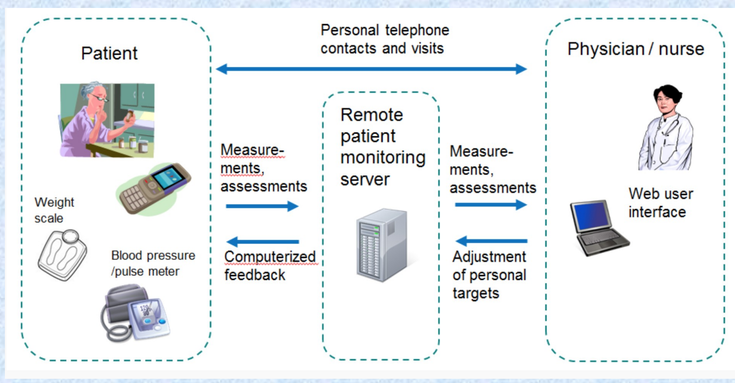
Patient	Physical functioning	Role Limit. Due to Phisical Problems	General Health	Vitality	Social Functioning	Role limit. Due to Emotional Problems	Mental Health	Pain
1	0	0	15	30	25	33	32	100
2	0	0	35	65	50	33	56	100
3	0	0	50	60	100	100	56	100
4	0	25	10	75	50	100	84	62

PIADS scale

Patient	Physical functioning	Role Limit. Due to Phisical Problems	General Health	Vitality	Social Functioning	Role limit. Due to Emotional Problems	Mental Health	Pain
1	0	0	15	60	50	100	52	100
2	0	0	35	70	75	100	72	100
3	0	0	50	60	100	100	70	100
4	0	25	10	75	50	100	84	62

Conclusions

- ✓ Our findings indicate that the system was user friendly for patients and care givers.
- ✓ Patient satisfaction scores were acceptable.
- ✓ The limitation of this study is that it has been applied only for a short period, yet. It is reasonable to suppose that a further evaluation after a long period could reveal more beneficial effects on patients and caregivers.
- ✓ The telemedicine system was effective for the home treatment of four siblings with FSHD and reduced the need for hospital admissions.



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